Studies from the Institute of Pathology

CASE VIII—A2512.

A Case of Mitral and Tricuspid Stenosis.

THE patient is a male, aged 24, whose family history reveals no relevant findings.

Previous Illnesses.

1. Eight years ago—meniscectomy following a knee injury.
2. Three years ago—attack of acute rheumatic fever.
3. Readmitted to Royal Victoria Hospital on five subsequent occasions with breathlessness on exertion, palpitation, and sense of fullness in head and epigastrium. These symptoms usually set in following a "cold" or overstrain.

History of present complaint.—The patient noticed a gradually increasing dyspnœa on exertion, with palpitation. Three days before admission the patient spat up some blood and complained of a pain in the right side of the chest. Since then he has noticed :—

1. Marked breathlessness even at rest.
2. Anorexia and insomnia.
3. Marked diminution of urinary output.
4. Swelling of the feet and ankles.
5. Feeling of heaviness below the right costal margin.
6. Headache.
7. Cough and spit, which is blood-stained.

The appetite was poor and bowels constipated, but moved readily with medicine. The amount of urine passed had diminished, and the urine was of a high colour. Sleep was poor.

On Examination.

The patient is a strongly-built male, showing some cyanosis of cheeks and ears. The skin is a lemon-yellow colour, which, with the cyanotic cheeks, has produced a very ashen-green colour of the facies. He is markedly orthopnœic. Gross œdema is present over the ankles and legs, and also over the sacrum. There is marked congestion of the external jugular veins, with venous pulsation to two and a half inches above the clavicle. Multiple small white punctate scars of an old infected chicken-pox rash are present over the body.

Alimentary system.—Tongue is heavily coated. Abdomen is distended, but shows bulging on both flanks—"burst tyre" effect. Shifting dullness and dullness in the flanks and both iliac fossæ can be elicited.

There is tenderness in the right costal margin, with an enlarged liver extending to the level of the umbilicus.

Cardio-vascular system.—Pulse-rate 104. Rhythm is regular, volume and tension are fair, and artery-wall is not palpable.

Blood-pressure 120/85.

Heart.—Marked generalised pulsation is present over the præcordium, with a palpable thrill at the apex. The apex beat is palpable in the fifth intercostal space five inches from the mid-line. D.A.C.D. = $\frac{iii}{1/5''}$

The heart-sounds are regular, but are almost completely obscured by a double "see-saw" type of murmur at the apex. The systolic element is rough and is conducted towards the posterior axillary fold. The diastolic element is softer and not conducted. This murmur can be heard all over the præcordium. In the pulmonary area the first sound is rumbling, and the second loud and slapping. No abnormal sounds can be heard in the aortic area or at Erb's point. In the tricuspid area the same "see-saw" murmur is present as occurred at the mitral area.

Respiratory system.—The chest expansion is poor, especially at the right base. Over the right base also, the lung resonance is impaired and the vocal fremitus and vocal resonance diminished. The breath-sounds are clear, except at the right base. Crepitations are present over both bases, but are considerably coarser on the right side.

W/R.—Negative.

Urine.—Acid. S.G. 1030. Trace of albumen. Urates + +.

X-ray chest.—There is considerable general enlargement of the heart, with a prominent pulmonary conus and dilated right auricle. Congestion is present at both lung-bases.

Electrocardiograph.—S_1 prominent. P. ii and iii prominent and bifid. Right ventricular preponderance.

Blood urea.—27 mgm. per cent. after admission. 43 mgm. per cent. shortly before death.

POST-MORTEM.

The body is that of a well-built young male, showing generalised icterus. The eyes and ears are cyanosed. Nail-beds are also blue, but there is no finger-clubbing. Gross œdema of lower limbs is present. Rigor mortis is commencing.

Serous cavities.—The peritoneal sac contains an excess of clear yellow serous fluid. The great omentum and appendices epiploicæ are a brick-red colour due to intense congestion. Both pleural sacs contain an excess serous transudate, which is most marked on the right side, while the pericardial sac contains about two ounces of a clear yellow fluid.

Heart.—The heart is grossly enlarged and globular in shape. The right auricle and great veins are dilated, and tense with blood. The tricuspid valve is the seat of a moderate stenosis, and the valve-cusps are shortened and thickened but show no recent vegetations. The right ventricle is enlarged and the wall hypertrophied. The endocardium of the left auricle is thickened and opaque, and there are calcified mural vegetations present in the posterior auricular wall. The mitral valve is grossly stenosed, and the valve-cusps hard and calcified. The left ventricle is not enlarged and the wall not hypertrophied. There are no changes in the aortic valve.

Left lung.—The lung feels heavy. The pleural surface is smooth. Small, hard nodules can be palpated throughout the lung substance. The bronchial mucosa is

intensely congested. On section the lung cuts firmly, and the changes of "brown induration" are well seen. On pressure a small amount of blood-stained fluid exudes, but there is no gross œdema. Scattered over the cut surface are a number of small hæmorrhagic infarcts of typical appearance.

Right lung.—The appearances are similar to the left, and the lung contains numerous infarcts, which are larger than those found in the left lung.

Liver.—This is enlarged and weighs four pounds. The surface is finely granular and presents some yellow-grey areas lying in a congested background. The liver cuts with some little difficulty. On section the nutmeg appearance of chronic venous congestion is well seen. Superimposed on the background is a distinct pattern of circumscribed yellow areas clearly marked off from the general background. Fibrosis is evident in relation to these areas and throughout the right lobe of the liver. The radicles of the portal vein and bile-duct appear normal. The gall-bladder contains no stones, and there is no obstruction to the biliary flow.

Spleen.—This is not much enlarged and weighs six ounces. The consistency is firm and the capsule smooth. On section it cuts firmly and the malpighian bodies stand out clearly, lying in a congested pulp.

Pancreas.—This is a little enlarged, and on section the acinar tissue appears normal. Scattered through the stroma of the gland and in nearby tissues are some yellow opaque areas of fat necrosis about the size of a pinhead.

Left kidney.—This kidney is enlarged and weighs eight ounces. The capsule strips readily, leaving a smooth surface. On section the organ is congested. The cortex is of normal width and the vascular pattern plain. The medullary striæ are prominent. There are no lesions in calyces, pelvis, or ureter.

Right kidney.—There is a double ureter to this kidney, connected to a double pelvis. The urine in the lower pelvis is turbid, but the mucosa shows no inflammatory changes. The appearance of this kidney otherwise does not materially differ from that of the left.

Urinary bladder.—The bladder appears healthy.

Alimentary tract.—Oesophagus: the mucosa is congested. Stomach: the mucosa is dark red in colour and of a velvety consistency. The surface is covered by a mucous secretion. There is no evidence of ulceration.

The rest of the alimentary tract shows some mild congestion of the mucosa, but no other lesions.

Adrenals.—These are of normal size, and the cortex is of average width and lipoid content. The medulla is grey in colour.

Thyreoid.—The thyreoid is a normal light brown colloid-containing gland.

Trachea.—The mucosa is intensely congested.

Aorta.—Shows no naked-eye changes.

Microscopical Examination.

Heart.—The mitral valve is thickened, fibrosed, calcified, and vascularised. There is general diffuse hypertrophy of the muscle fibres throughout the heart. Paravascular fibrous scars with a characteristic fibrillar appearance are scattered throughout the stroma of the heart.

Aorta.—None of the coats of the aorta shows any specific changes.

Lungs.—The typical microscopical changes of C.V.C.* are present, with thickened fibrosed alveolar walls, prominent alveolar wall capillaries, and alveoli packed with red blood-cells, and hæmosiderin containing macrophages (heart-failure cells). Throughout the lung the smaller branches of the pulmonary artery show evidence of a thickened intima, which is produced by a proliferation of the subendothelial connective tissue.

Superimposed on this basis of chronic passive congestion and diseased vessels there are numerous scattered hæmorrhagic infarcts of normal histological appearance, many of which are of some duration and show necrotic changes in the centre.

Liver.—The pathological changes in the liver vary greatly from one area to the next. There is marked chronic passive congestion present, with a blood-lake in the central part of the liver lobule in some areas. Some of the lobules show fibrosis and reticular collapse in the mid-zone of the lobules, while areas of liver-cell hyperplasia are present with bile-duct proliferation. The whole makes up a composite picture of chronic passive congestion, liver-cell damage, liver regeneration, and fibrotic replacement. Such appearances are typical of a long-standing chronic passive congestion, with periods of recovery, leading to a "cardiac cirrhosis" of the liver.

Kidneys.—The kidneys, apart from the marked venous congestion, are remarkably healthy and show no microscopical changes beyond a little albuminous fluid in Bowman's capsule and in some of the tubules.

Urinary bladder.—The mucous membrane is desquamated and the submucosa fibrosed with prominent blood-vessels.

Spleen.—The malpighian bodies are clearly marked off from a congested pulp, in which sinusoids are dilated with blood and show extensive fibrosis of their walls.

Pancreas.—There is a mild degree of post-mortem autolysis in the acinar and islet tissue.

Adrenals.—The cortex shows some loss of the cortical lipoid with a marked increase of the fibrous stroma. Focal areas of round-celled infiltration are present in the medulla.

Other organs.—The other organs showed no significant microscopical changes.

Anatomical diagnosis.

Mitral stenosis : calcification of cusps.
Hypertrophy of right ventricle : tricuspid stenosis.
Healed rheumatic carditis.
Gross C.V.C. of liver, lungs, spleen, and kidneys.
Cardiac cirrhosis.
Infarcts in lungs.
Pulmonary endarteritis fibrosa.

*Chronic venous congestion.

Commentary.

Congestive heart failure is, unfortunately, quite a common clinical condition, and it is usually the end result of an unsuccessful battle waged between the heart muscle and some mechanical difficulty to the normal blood flow. This difficulty to the blood flow often takes the form of some valvular damage to the heart, either a valvular insufficiency or stenosis. Thus, when such a patient dies he will show pathological changes, due to two causes :—

1. Changes produced by the mechanical insufficiency in the circulation.
2. Changes caused by the etiological agent which has damaged or is damaging the heart-valves.

The changes produced by either of these factors will vary in degree and extent, and it is unusual for both to be equally prominent in any one case.

The clinical course followed in this case is in many ways typical of that group of cases of congestive heart failure, in which the etiological factor is of rheumatic origin. However, certain abnormal clinical features presented interesting problems which could best be solved in a clinico-pathological correlation.

The patient lived a normal, healthy life until he was twenty-one, when he developed an attack of acute rheumatic fever, which caused the initial cardiac damage. Subsequent recrudescences of the rheumatic infection, induced by intercurrent infections which were mainly of the upper respiratory tract, produced further cardiac damage. The brunt of the rheumatic infection fell on the myocardium and on the mitral valve. During the recrudescences of the infection the myocardium suffered further insults, while the mitral valve cusps became thickened, fibrosed, shortened, and at last calcified. Stenosis of an extreme degree resulted.

Evidence of involvement of the tricuspid valve with stenosis was present, but the changes were less marked and less advanced than in the mitral valve and were probably of not so long standing. During some of these later relapses, signs of congestive heart failure became evident and indicated the extremely low state of the myocardial reserve. Under medical treatment the patient's clinical condition improved, and on each occasion after treatment the patient was discharged from hospital much improved, but he had only to return each time intercurrent infection or overstrain again reduced the cardiac reserve to zero.

The rheumatic nature of the lesion described in the clinical history was borne out by the pathological examination. Stigmata of previous rheumatic carditis were present in the shape of typical valvular and myocardial lesions. Histological examination revealed the presence of para-vascular fibrillar fibrous scars scattered throughout the myocardium, which resembled in all detail the descriptions given by Gross of healed Aschoff nodes. No recent Aschoff nodes were seen, so that active rheumatic infection was absent at the time of the patient's death, which must have been due to mechanical causes dictated by an inefficient valvular apparatus and a myocardial reserve strained to the uttermost.

The case, in its final state, was therefore an example of combined mitral and

tricuspid stenosis. Such multiple valvular lesions present many interesting problems, both in diagnosis and hæmodynamics. This case was no exception.

The ante-mortem diagnosis of acquired tricuspid stenosis has been rarely made. Of the two hundred and fifty cases recorded in the literature, an ante-mortem diagnosis has only been made in thirty-one. There are many reasons, apart from the rarity of the condition, which account for this.

The condition is rarely found alone, and the commonest condition with which it is associated is mitral stenosis. This greatly complicates the detection of tricuspid stenosis, as the signs of mitral stenosis dominate the picture and obscure those produced by the tricuspid valve.

Relative tricuspid insufficiency, which is said to accompany frequently long-standing mitral stenosis, also presents considerable difficulty.

However, from the pathological physiology the following clinical manifestations have been deduced, which indicate the presence of a tricuspid stenosis :—

1. Dyspnœa on exertion, out of proportion to that found in mitral stenosis.
2. Cyanosis, often of extreme degree.
3. Distention of the cervical and brachial veins, with marked presystolic pulsation due to right auricular contraction.
4. Enlargement of the liver, with presystolic pulsation due to the same cause.
5. A diastolic or presystolic murmur heard best at the ziphoid, which usually differs in quality but not in timing from the murmur at the apex.
6. Enlargement of the heart to the right, with dilatation and hypertrophy of the right auricle.
7. The signs dependent on auricular activity will disappear during auricular fibrillation, auricular arrest, etc.

A diagnosis of organic tricuspid stenosis is justified if a patient with a chronic rheumatic heart and mitral stenosis presents the above signs. One must first exclude mitral stenosis and adhesive pericarditis, and mitral stenosis and patent foramen ovale, which are rare conditions presenting similar signs.

Pathologically, tricuspid stenosis presents no new features. The valve undergoes the same changes as the mitral and ends in healing. The right auricle is usually distended and its wall hypertrophied, and in one case reported the right auricle actually held 2,500 cc. of water. The right auricular enlargement is evident on both clinical and X-ray examination by enlargement of the heart to the right. A grossly enlarged heart on X-ray examination is not a common finding in pure mitral stenosis.

The changes found in the liver in this case were both unusual and interesting. They present the pathological features of that condition known as "cardiac cirrhosis," which has been much stressed in the French literature, but has received scant attention here. The condition was first described by Becqueruel in 1840, who suggested that C.V.C. from cardiac failure could produce hepatic cirrhosis—cirrhose cardiaque. Later it was produced experimentally by mechanically obstructing the inferior vena cava. Roussy pointed out in 1853 that when C.V.C. was prolonged, the picture of C.V.C. of the liver became complicated by regeneration of hepatic

tissue to form hyperplastic nodules and associated proliferation of connective tissue in two sites : (a) around the central vein, (b) around hypertrophic islands. Such a liver, on naked-eye examination, appears irregularly granular on the surface and shows on section pale yellow areas separated clearly from a background of a congested and cirrhosed liver.

There are no pathognomonic clinical signs which indicate the presence of cardiac cirrhosis, and jaundice, which occurs in ten per cent. of all cases of congestive heart failure, is not a constant clinical finding.

The only suggestive evidence is purely of an inferential nature and is given by the clinical history. This is the presence of severe recurrent episodes of congesitve heart-failure over a long period, with intervals of improvement in cardiac function, when the liver recovers from the venous stasis and regeneration takes place. However, even with such favourable conditions, cardiac cirrhosis does not always result.

The pathogenesis of the condition is readily explained. The degeneration of the liver cells resulting from the C.V.C. varies in extent and in severity in different lobules. The damage to the liver epithelial cells occurs during the phase of cardiac failure. Those lobules which have only been partially destroyed are repaired during periods of improvement in the cardiac function. When entire lobules are destroyed the change is irreversible, the supporting reticulum collapses, and fibrous tissue proliferation replaces the area by a fibrous scar.

The naked-eye and microscopical lesions in the liver of this case all tend to confirm that the liver is the seat of an advanced cardiac cirrhosis.

The lungs showed the changes of C.V.C. as the result of the long-standing mitral stenosis. This was clearly manifested by the "brown induration" of the lungs and the fibrosis of the alveolar walls, with the presence of red blood-cells and "heart-failure cells" in the lung alveoli. A small but persistent hæmoptysis was clinical evidence of this C.V.C. of the lungs.

The final illness was ushered in by pain in the chest, cough, and hæmoptysis. Clinical and post-mortem examination confirmed the presence of lung infarcts. Careful search was therefore made in the right auricle and auricular appendage for the presence of any thrombus which might have become partially dislodged, to impact in one of the branches of the pulmonary artery to produce the infarcts in the lungs. None, however, was found. Although the possibility of a thrombus having become completely dislodged without leaving any trace was considered, it was thought that in all probability the thrombus was locally formed in the lung-vessels. The presence of long-standing pulmonary hypertension was known to have been existent, and reactive changes with intimal hyperplasia was evident in the pulmonary vessels. Secondary degenerative changes occurring in the intima of such vessels could readily be the nidus for local thrombosis, especially with a reduction in the normal blood-flow induced by heart failure. This local thrombus formation, with subsequent lung infarction, might explain many cases of lung infarcts occurring in mitral stenosis, where the presence of a source for embolism is non-existent.

The inadequacy of the general systemic circulation resulted in anoxæmia, reduced

capillary blood-flow, raised venous pressure, and, because of the liver damage, some reduction in the plasma proteins is probable. Because of these factors peripheral œdema was marked, and generalised anasarca present. Portal hypertension, the result of the heart failure and some obstruction to the portal circulation in the liver, was shown by ascites, marked congestion of the mesenteric vessels, and a congested firm spleen. The spleen showed the typical changes of long-standing C.V.C. with fibrosis of the splenic sinusoidal walls.

The congestion of the kidneys, the pre-renal deviation of fluid to œdematous tissues, and the inadequate renal blood-flow were all reflected on examination of the urine. Oliguria was present, with the passage of small amounts of a high S.G. urine containing albumen and urates. The kidneys responded well to the use of mercurial diuretics and the renal excretion markedly improved, with a concomitant reduction in the œdema. At this time the kidneys showed no difficulty in ridding the body of unwanted waste products, and the blood urea was normal. However, as the cardiac condition deteriorated and the rate of blood-flow fell, the dependence of the kidney on the general state of the cardio-vascular system became evident. The output, therefore, diminished in spite of diuretics, and the blood urea rose shortly before death.

Death occurred after a long illness in an unavailing fight between the heart and an inadequately adjusted circulatory system which ended in marked right heart failure, showing gross œdema, C.V.C. of all organs, and fluid in all the serous sacs.

SUMMARY.

1. A case of rheumatic heart disease showing mitral and tricuspid stenosis is presented.
2. Points in the ante-mortem diagnosis of acquired tricuspid stenosis are made.
3. The liver showed the changes of "cardiac cirrhosis" which is discussed.

M. G. N.

REVIEWS

ELEMENTARY PATHOLOGICAL HISTOLOGY. By W. G. Bernard, F.R.C.P. (Lond.). Second Edition. 1940. London: H. K. Lewis & Co., Ltd. Pp. 75. Plates 181, Coloured 8. Price 10/-.

If every medical student could be induced to purchase this book, and keep it by his side on the bench when examining sections, he would quickly gain a sound basic knowledge of pathological changes involved in diseased tissues, instead of, as so often happens, a facility in merely "spotting sections." The book consists mainly of 181 micrographs of typical pathological conditions (eight of which are in colour), with a concise but accurate account of the changes from the normal in each condition. These short descriptions attempt to interest the student to try and resolve his own particular section into the various cells, particles, and substances of which it is composed, and thus to learn to analyse the changes represented from the normal, and to add them into a coherent account of what has happened to the tissue. It is only by such means that a true appreciation of pathology can be obtained, and it is only on such a true appreciation that a sound knowledge of medicine can be built. It is therefore a pleasure to strongly commend this book to the attention not only of undergraduates but to those who, while being beyond that stage, remain students in the true sense.